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ORIGINAL RESEARCH

Comparison of respiratory muscle strength, quality of life and functional capacity among adolescents with cystic fibrosis with different bacteriological profiles

Comparação da força muscular respiratória, qualidade de vida e capacidade funcional entre adolescentes com fibrose cística com diferentes perfis bacteriológicos

Comparación de la fuerza muscular respiratoria, la cualidad de vida y la capacidad funcional entre adolescentes con fibrosis quística con distintos perfiles bacteriológicos

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ABSTRACT | We compared the respiratory muscle strength, quality of life and functional capacity in adolescents with cystic fibrosis with different bacteriological profiles. This is a cross-sectional study of descriptive character on adolescents with cystic fibrosis assessed in the Reference Center for Cystic Fibrosis of the Octávio Mangabeira Specialized Hospital (HEOM) from January to March 2016, characterizing a sample for convenience. Muscle strength evaluation with the manovacuometer was performed, as well as analysis of the quality of life through the application of quality-of-life questionnaire with validation for patients with cystic fibrosis (CFQ), analysis of the functional capacity, after the six-minute walk test, and bacteriological profile, through results of microbiological tests. We evaluated 30 individuals with cystic fibrosis, in which, for *Staphylococcus aureus*, respiratory muscle strength (75.6±19.6*), quality of life CFQ (59.3±3.4), and functional capacity (427.8±64.6*) were verified. For *Pseudomonas aeruginosa,* the respiratory muscle strength (61.4±19.1*), quality of life CFQ (47.9±4.2) and the functional capacity (382.0±78.0*) were analyzed. We concluded that both bacteriological profiles impair lung function, especially the bacterium *Pseudomonas aeruginosa,* which has a tendency to respiratory muscle weakness, especially for women (Mip<60cmH2O) and low functional capacity.

Keywords | Cystic Fibrosis; Muscle Strength; Exercise Tolerance; Quality of life.

RESUMO | Comparou-se a força muscular respiratória, qualidade de vida e capacidade funcional em adolescentes com fibrose cística com diferentes perfis bacteriológicos. Trata-se de um estudo transversal de caráter descritivo, em adolescentes com fibrose cística avaliados no Centro de Referência em Fibrose Cística do Hospital Especializado Octávio Mangabeira (HEOM) no período de janeiro a março de 2016, caracterizando uma amostra por conveniência. Foi realizada avaliação da força muscular com o manovacuômetro, análise da qualidade de vida por meio da aplicação do questionário de qualidade de vida com validação para pacientes com fibrose cística (QFC), análise da capacidade funcional, após a realização do teste de caminhada de seis minutos, e do perfil bacteriológico, por intermédio de resultados de exames

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de microbiologia. Foram avaliados 30 indivíduos com fibrose cística, em que, para *Staphylococcus aureus*, verificaram-se a força muscular respiratória (75,6±19,6*), a qualidade de vida QFC (59,3±3,4) e a capacidade funcional (427,8±64,6*). Para *Pseudomonas aeruginosa* foram analisadas a força muscular respiratória (61,4±19,1*), a qualidade de vida QFC (47,9±4,2) e a capacidade funcional (382,0±78,0*). Concluiu-se que ambos os perfis bacteriológicos comprometem a função pulmonar com ênfase para a bactéria *Pseudomonas aeruginosa*, que apresenta tendência à fraqueza muscular respiratória, principalmente para o sexo feminino (PImáx<60cmH2O) e baixa capacidade funcional.
Descritores | Fibrose Cística; Força Muscular; Tolerância ao Exercício; Qualidade de Vida.

RESUMEN | Se comparó la fuerza muscular respiratoria, la cualidad de vida y la capacidad funcional en adolescentes con fibrosis quística con distintos perfiles bacteriológicos. Se trata de un estudio transversal de carácter descriptivo, en adolescentes con fibrosis quística que fueron evaluados en el Centro de Referencia en Fibrosis Quística del Hospital Especializado Octávio Mangabeira (HEOM) en el período de enero a marzo de 2016, caracterizando una muestra por conveniencia. Fue realizada la evaluación de la fuerza muscular con el manovacuómetro, el análisis de la cualidad de vida por medio de la aplicación del cuestionario de cualidad de vida con validación para los pacientes con fibrosis quística (CFQ), el análisis de la capacidad funcional, después de la realización de la prueba de caminata de seis minutos, y del perfil bacteriológico, por intermedio de resultados de análisis de microbiología. Fueron evaluados a 30 individuos con fibrosis quística, en que, para *Staphylococcus aureus*, se certificaron la fuerza muscular respiratoria (75,6±19,6*), la cualidad de vida CFQ (59,3±3,4) y la capacidad funcional (427,8±64,6*). Para *Pseudomonas aeruginosa* fueron analizadas la fuerza muscular respiratoria (61,4±19,1*), la cualidad de vida CFQ (47,9±4,2) y la capacidad funcional (382,0±78,0*). Se concluyó que ambos perfiles bacteriológicos comprometen la función pulmonar con énfasis para la bacteria *Pseudomonas aeruginosa*, que presenta tendencia a la debilidad muscular respiratoria, principalmente para el sexo femenino (PImáx<60cmH2O) y baja capacidad funcional.
Palabras clave | Fibrosis Quística; Fuerza Muscular; Tolerancia al Ejercicio; Cualidad de Vida.

INTRODUCTION

Cystic fibrosis (CF) is an inherited and progressive disease associated with the deterioration of lung function, malnutrition and progressive limitation to exercise[1]. Its incidence varies according to ethnic groups, affecting approximately 1 every 2,500 white children born alive and, proportionally, 1 to 17 thousand black individuals. In Brazil, it is estimated at 1/10,000 live births[2].

Respiratory changes in CF consist of pneumothorax, hemoptysis, nasal polyps and chronic obstructive pulmonary disease, whose progression is the main cause of morbimortality in patients with cystic fibrosis[2-4]. CF is marked by inflammation in the airways and the most relevant lung changes are obstruction of the airflow, air trapping and inadequate ventilation. The pattern of evolution of these changes is characterized by prevalence of obstructive ventilatory disorder with early reduction of flows related to the small airways and late impairment of forced vital capacity (FVC)[5].

The impairment of pulmonary function is characterized by respiratory infection followed by colonization by bacteria. In this situation, the infectious process increases the obstructive phenomenon, resulting in a vicious circle, difficult to be stopped. Although the obstructive process is the initial pathophysiological event, the chronic infection of the respiratory tract is the most important event, thus contributing to the worsening of pulmonary function[6] and of functional capacity. Recent findings suggest that the reduction in the distance walked in the six-minute walk test (6MWT) is associated with the decreased functional ability[6].

Due to functional repercussions, the evaluation of respiratory muscle strength is an important outcome in patients with CF and essential parameter in clinical practice, since these muscles are responsible for the proper functioning of the respiratory system[7].

Thus, the overall objective of the study was to compare the respiratory muscle strength, quality of life and functional capacity in adolescents with cystic fibrosis with different bacteriological profiles.

METHODOLOGY

This is a cross-sectional study of descriptive character, carried out from January to March 2016. It was approved by the Research Ethics Committee

involving humans of the Instituto de Ciências da Saúde at the Universidade Federal da Bahia (ICS-UFBA) under the protocol number 1,377,967.

Participants

The convenience sample consisted of 30 patients with CF clinical diagnosis, who attended the Reference Center for Cystic Fibrosis of the Octávio Mangabeira Specialized Hospital (HEOM), located in the city of Salvador, Bahia. We included both sexes, aged between 12 and 18 years, diagnosed with cystic fibrosis, participants of the physical therapy program, who signed the Informed Consent Form (ICF) and the Free and Clarified Consent Term. Patients with orthopedic limitations, diagnosis of respiratory diseases other than CF, cardiovascular diseases, and cognitive deficit were excluded.

Materials and procedures

After sample selection, we recorded demographic data on age, sex, race, body mass index (BMI), bacteriological profile and anthropometric weight measurements using a digital scale (Magna 150 kg, G-Life, CA4000), and height, with metric tape of 150 cm and meterage on both sides.

BMI was calculated from the weight in kilograms divided by the square of height in meters, being considered a useful and efficient measure in the evaluation of excess body fat. For individuals aged up to 20 years, we recommend the BMI assessment with the aid of reference curves. The indicator is assessed using cut-off points, being considered low weight the individual who is below 5 %, eutrophic between 5 and 85 %, overweight if the BMI is greater than or equal to 85 %, and obese above 95 %[8,9].

The evaluation of the maximum respiratory pressure occurred through an analog manovacuometer (WIKA, model 611.10), whose scale presents intervals of 10cmH2O and variation from –150 to +150cmH2O. To measure the maximum inspiratory pressure (Mip), the subject was sat and instructed to breathe out slowly. When in residual volume, a mouthpiece was connected between their lips, closing both the nasal cavity with a clip and, from then on, an intense and deep inhalation was produced in the manovacuometer. To measure the maximum expiratory pressure (Mep), the subject, still in the sitting position, inhaled slowly until the total lung capacity (TLC) and then compressed the oral adapter between the lips and the nasal airway was occluded with the clip, so that the subject could inhale fast and intensely in the manovacuometry tool. Both pressures were measured three times, interspersed with 2 minutes rest, considering only the highest value, but all measurements were recorded. Between Mip and Mep measurements, there was a range of five minutes[10]. The values specified for Mip and Mep in adolescents were obtained from the reference equation: Female Mip: –33.854–(1.814×age); Male mip: –27.020–(4.132×age); Female Mep: 17.066+(7.22×age); Male Mep: 7.619+(7.806×age)[11].

We applied a quality-of-life questionnaire with validation for patients with cystic fibrosis (CFQ). The questionnaire used in this study had two versions: the CFQ_12-13 years (35 questions) and the CFQ_over 14 years (50 questions). This instrument assesses the following domains: physical, body image, emotional, social/school, vitality, food, treatments, digestive, respiratory, weight and health. The scores of each domain range from 0 to 100, the value 100 refers to very good QoL and, generally, score from 50 to 100 reflects good QoL. CFQ has been applied in the form of interview, regardless of the age group. Respondents were encouraged to respond according to their experiences, information and personal opinions, clearing up their doubts, if necessary. The interviews, conducted always by the same researcher, lasted, in total, between 20 and 30 minutes.

Then, the evaluator recorded the heart rate values (HR, bpm) and peripheral oxygen saturation (SpO2, %) of the subject through a portable pulse oximeter (*Contec* MED CMS-50D, Commercial Society), blood pressure (BP, mmHg), by using an automatic arm blood pressure device (OMRON, model HEM-7113), respiratory rate (RR, ipm), and the degree of dyspnea and fatigue by modified Borg scale. The measurement of vital data, such as HR, RR, blood pressure (BP), oxygen saturation (SpO2) and degree of dyspnea through the Borg scale are suggested by Rodrigues et al.[12] before, at the end of the six minutes and three minutes after its termination. After assessing the vital data, the subject was directed to a hallway with flat floor, 30 m long, with markings every 3 m, in open environment, in order to carry out the six-minute walk test (6MWT). Before the test, the subject received guidance to walk quickly, not being allowed to jump or run and, if he/she felt some discomfort, the 6MWT could be stopped, but the test time would still

be counted. The researcher remained standing with the stopwatch on one end of the hiking trail, signaling every minute the time remaining until the end of the test, offering verbal encouragement in a neutral tone ("you are doing great" and "keep at it"). After six minutes, the subject was instructed to stop, and the distance walked from the starting point until the stopping point was measured. The predicted values for the distance walked in the 6MWT in adolescents were obtained from the reference equation: Female: DW6m = 217.06 + (0.6 × Age years) – (2.04 × Weight Kg) + (3.24 × Height cm); and Male: DW6m=145.21+(4.62×Age years) – (3.84 × Weight Kg) + (4.01 × Height cm)[13].

Bacteriological profile for *Staphylococcus aureus* or *Pseudomonas aeruginosa* has been set from the result of microbiological tests (sputum culture), of the last six months, being considered the most recent sample. Sputum culture was conducted following the Protocol of HEOM, preferably in the morning, by a proper professional and through the expectoration in sterile container, properly identified.

Statistical analysis

The statistical analysis was catalogued in Microsoft Office Excel 2007 and the variables were analyzed by the statistical software SPSS version 20.0. To test the normality of the sample distribution, we used the Shapiro-Wilk test. Continuous variables with normal distribution were expressed as mean and standard deviation, while the nominal and ordinal variables were expressed as absolute and relative frequency. The Student's t test was performed for comparison of variables and the level of significance established was $p<0.05$.

The Spearmann test was performed to correlate LCADL and SGRQ, expressed by the value "r." We considered: intensity of small correlation ("r" until 0.25), low ("r" between 0.26-0.49), moderate ("r" between 0.50-0.69), high ("r" between 0.70-0.89) and very high ("r" above 0.90), according to Gross et al.[14], considering $p<0.05$ as statistically significant difference.

RESULTS

We evaluated 30 subjects, whose demographic data are described in Table 1.

Table 1.

Sociodemographic data	n=30
Age, years	14.4±1.8
Sex, male/female	17/13
White/black	9/21
BMI, kg/m²	
Underweight (below 5 percentile)	5 (14.9±0.9)
Appropriate weight (between 5-85 percentile)	20 (18.1±2.0)
Appropriate weight (between 5-85 percentile)	2 (24.1±0.6)
Obesity (above 95 percentile)	3 (27.3±4.2)
BP	
Staphylococcus aureus/Pseudomonas aeruginosa	16/14
CFQ_{dt}	53.9±6.9

BMI: body mass index; BP: bacteriological profile; CFQ_{dt}: total sum of quality-of-life questionnaires. Results expressed in number and (mean±standard deviation)

No statistically significant difference was found for the variable Mep on the comparison of values obtained for the respiratory pressure and the distance walked in the 6MWT for demographic data on adolescents. However, statistically significant difference was found for the variable Mip between sexes (p=0.03), for the groups underweight and appropriate weight (p=0.005), and for the bacteriological profile group, *Staphylococcus* (p=0.04), *Pseudomonas* (p=0.03). The result was also statistically significant to the variable distance walked in the 6MWT between the race group (p=0.03) and the group bacteriological profile *Staphylococcus* (p=0.03), *Pseudomonas* (p=0.02), as shown in Table 2.

Table 2. Values of maximum respiratory pressure and the distance walked in the 6MWT of patients with cystic fibrosis

	Mip (cmH_2O)	Mep (cmH_2O)	DW (m)
Sex			
Male	75.8±19.7*	64.1±9.4	426.3±60.2
Predicted	86.5	120.3	436.4
Female	60.0±18.2*	61.5±14.6	380.4±83.6
Predicted	75.5	120.0	402.3
Race			
White	61.1±18.3	60.0±10.0	362.6±65.2*
Black	72.3±20.7	64.3±12.4	425.2±70.3*
BMI, kg/m²			
Underweight	48.0±13.0*	58.0±13.0	362.8±85.5
Appropriate weight	74.5±18.7*	63.5±11.3*	411.6±67.5
Overweight	65.0±7.1	60.0±7.1*	439.5±102.5*
Obesity	70.0±30.0*	70.0±17.3	422.6±95.5*
BP			
Staphylococcus	75.6±19.6*	70.0±17.3	427.8±64.6*
Pseudomonas	61.4±19.1*	61.4±10.2*	382.0±78.0*

Mip: maximum inspiratory pressure; Mep: maximum expiratory pressure; DW: distance walked in the 6MWT; BMI: body mass index; BP: bacteriological profile; *P<0.05. Results expressed in mean±standard deviation

Table 3 shows the measured values of the CFQ domains for bacteriological profile, *Staphylococcus aureus* and *Pseudomonas aeruginosa*, statistically significant

difference was found for *Staphylococcus aureus* in the physical (p=0.02), emotional (p=0.02), social/school (p=0.04), food (p=0.01), respiratory (p=0.01) and health (p=0.02) domains; and for *Pseudomonas aeruginosa* in the physical (p=0.01), emotional (p=0.002), social/school (p=0.04), food (p=0.01), respiratory (p=0.01), weight (p=0.04) and health (p=0.01) domains.

Table 3. Values of the domains of the questionnaire validated for cystic fibrosis (CFQ) for bacteri*ological profile, Staphylococcus aureus and Pseudomonas aeruginosa*

CFQ Domains	SPA	PA
Physical	66.6±10.6*	49.0±7.07*
Body image	62.6±10.4*	55.4±11.6*
Emotional	64.1±13.2*	52.5±13.3*
Social/school	64.2±10.2*	51.9±11.0*
Vitality	62.0±12.5	49.7±9.9
Food	51.7±11.5*	40.8±10.6*
Treatments	61.8±14.0	54.4±10.8*
Digestive	55.0±11.8	47.6±12.0
Respiratory	51.3±8.4*	34.0±8.5*
Weight	48.1±11.2	37.3±8.7*
Health	54.4±10.5*	41.5±7.1*
CFQ_{dt}	59.3±3.4	47.9±4.2

SPA: *Staphylococcus aureus*; PA: *Pseudomonas aeruginosa*; CFQ_{dt}: total sum of domains in the quality-of-life questionnaire. *P<0.05. Results expressed in mean±standard deviation

Table 4 shows the correlations of Bacteriological profiles *Staphylococcus aureus* and *Pseudomonas aeruginosa* with the variables of maximal respiratory pressures, distance walked in the 6MWT and questionnaire validated for cystic fibrosis (CFQ). The correlations between EP1 and DW1, IP1 and EP1, and IP2 and DW2 showed significant correlation at the level 0.01. The correlations between EP2 and DW2, EP2 and CFQ2, IP1 and DW1, and IP2 and CFQ2 showed significant correlation at the level 0.05.

Table 4. Correlations of bacteriological profiles *Staphylococcus aureus* and *Pseudomonas aeruginosa* with the variables of maximal respiratory pressures, distance walked in the 6MWT and questionnaire validated for cystic fibrosis (CFQ)

	IP2	EP2	DW2	CFQ2	IP1	EP1	DW1	CFQ1
IP2	1	0.458	0.802**	0.649*	0.215	0.057	0.107	0.166
EP2		1	0.559*	0.590*	−0.3	−0.27	−0.21	0.39
DW2			1	0.053	0.011	−0.09	−0.01	0.05
CFQ2				1	−0.21	0.052	−0.03	0.356
IP1					1	0.724**	0.515*	0.071
EP1						1	0.679**	−0.02
DW1							1	−0.09
CFQ1								1

1: *Staphylococcus aureus*; 2: *Pseudomonas aeruginosa*; IP: maximum inspiratory pressure; EP: maximum expiratory pressure; DW: distance walked in the 6MWT; CFQ: questionnaire validated for cystic fibrosis. **The correlation is significant at the level 0.01. *The correlation is significant at the level 0.05

DISCUSSION

Most individuals in the sample presented deficiency of respiratory muscle strength obtained from the measurement of Mip and Mep, both in cmH2O. The measurement of these characteristics enables the achievement of significant indexes for the functional evaluation of respiratory muscles[10].

We verified that men and women with cystic fibrosis showed Mip and Mep below the reference value calculated for this population. The predicted values were obtained from the reference equation for adolescents described by Domènech et al.[11].

A high Mip (>80cmH2O) or a high Mep (>90cmH2O) exclude, respectively, the inspiratory or expiratory weakness. To indicate whether a person has a low Mip, this value must be less than 60% of the predicted value, which is based on variables such as sex and age, body weight and height; the values above 60cmH2O exclude clinically the weakness of respiratory muscles[10]. In our study, women showed a Mip average of 60.0±18.2, characterizing the group with weak respiratory muscle strength. In the individual analysis of the sample, 11 individuals showed Mip below or equal to 60cmH_2O.

Specific questionnaires such as the CFQ are focusing on the signs and symptoms of a single disease, such as cystic fibrosis, which allows to evaluate how the disease affects the daily functions[14]. The results of Ribeiro et al.[15] shows that children aged between 6 and 14 years have a satisfactory QoL in CFQ. However, adolescents and adults with increased pulmonary impairment showed worse scores in respiratory, social role, body image and physical domains. This study showed a total domain average of 53.97±6.9 for CFQ, which represents good quality of life for these individuals. We considered the physical, respiratory, treatments, social/school, food, health and weight domains as statistically significant in the analysis between groups, showing, overall, relevant impairment to *Pseudomonas aeruginosa* (p=0.03). Ribeiro et al.[15] considers important the QoL assessment of those individuals, since it corresponds to the patient's perception about what it is like to live with a chronic disease.

As for functional capacity, the gradual reduction of the physical conditioning linked to inactivity starts a vicious cycle in which the worsening of dyspnea is related to increasingly small physical efforts, with serious quality of life impairment[16]. Thus, the 6MWT is

being a test widely used to assess the functional capacity of the individual with chronic degenerative diseases, as well as in cystic fibrosis, estimating the patient's tolerance of daily life activities. Corroborating our findings, Coelho et al.[17] showed in their study that children and adolescents with cystic fibrosis present decreased aerobic performance and, consequently, reduced distance walked due to alteration in the lung function.

Enright and Sherrill[18] reported that the distance walked in the 6MWT is an important predictor of morbidity and mortality in patients with lung disease and/or cardiovascular disease, especially those who walked a distance shorter than 300 m. Although some subjects of our study present test values below the reference value, all these individuals walked more than 300 m at the end of the 6MWT, showing that, in our sample, the patients did not present higher risk of morbidity and mortality for cystic fibrosis.

In both sexes, the distance walked in the 6MWT was below the reference value for those patients with cystic fibrosis, resulting in low functional capacity. As well as in our research, Ziegler et al.[19] found that 73.2% of the 41 adolescents and adults with cystic fibrosis that compose their sample walked a distance below the lower limit of normal in the 6MWT. However, different from our study, Chetta et al.[20] analyzed 25 adult patients (15 women aged 18-39 years) diagnosed with cystic fibrosis, showing a normal exercise capacity in relation to the distance walked in the 6MWT. Oliveira[13] describes the reference value to the distance predicted in the 6MWT for adolescents.

The measurement of vital data, such as HR, RR, PA, SpO2 and degree of dyspnea through the Borg scale is suggested by Rodrigues et al.[12] before, at the end of the six minutes and three minutes after they finished. Studies with healthy children show that oxygen saturation varies little during testing, while elevations in the cardiac and respiratory frequencies are highlighted. Our findings are in accordance with the one presented by Lammers et al.[21], adding that these variables returned to resting values a few minutes after the test.

The bronchial tree colonization by agents such as *Pseudomonas aeruginosa* and/or *Staphylococcus aureus* has suggested serious impact on pulmonary function. According to Magalhães et al.[22], chronic inflammation, secondary to the infectious process, causes irreversible damage in the bronchial walls, determining the degradation with consequent decline in lung function.

According to the study of Reis et al.[23], *Pseudomonas aeruginosa* has been the most important bacteria of the respiratory tract of patients with cystic fibrosis, with colonization rates ranging from 50 to 70%, in different treatment centers. Although patients may be colonized in the first years of life, the microorganism is not commonly isolated from the respiratory tract until late childhood and early teens, following colonization by *Staphylococcus aureus*. In our study, there was the presence of colonization by both bacteria, with higher prevalence in the study population affected by *Staphylococcus aureus*, but with the greatest impact for individuals affected by *Pseudomonas aeruginosa*.

The *Pseudomonas aeruginosa* bacterium is the main responsible for the progressive damage and relevant impact on pulmonary function. The incidence of *Pseudomonas aeruginosa* increases with age and reaches 70 to 90% of the patients[24]. In our findings, the presence of this bacterium presented statistically significant data for lung function deficiency (p=0.03).

Corroborating our findings, in which the *Pseudomonas aeruginosa* presented important impairment in the weight (p=0.01) and food (p=0.01) domains in CFQ and overall relevance in QoL, Lammers et al.[21] claim that the pulmonary disease process leads to increased energy expenditure, aggravating the nutritional status in cystic fibrosis, especially before a chronic infectious process, as in the case of *Pseudomonas aeruginosa* and of the progressive disease with airflow obstruction, which may aggravate these patients' quality of life.

As to the sample size, some important aspects may have contributed to the relatively small population found in the study, among them: the population restricted to adolescents, some of them from the interior of Bahia, show transportation difficulties and considerable absence in school due to treatments.

We can also point out that the sample calculation was not performed, because there was no prior or pilot study in the literature, characterizing thus a study limitation.

We can consider the existence of several gaps in the literature on individuals with cystic fibrosis, especially concerning adolescents. In addition, material linking the bacteriological profile with respiratory muscle strength, quality of life and functional capacity was not found. Therefore, this study is unprecedented, though with some limitations, especially regarding the theoretical background.

CONCLUSION

In this context, we concluded that both bacteriological profiles impair lung function, especially the bacterium *Pseudomonas aeruginosa*, representing a greater tendency to respiratory muscle weakness in this group, mainly for women (Mip<60cmH2O). Still, one can infer that individuals presented low functional capacity, because they walked a distance in the 6MWT below the one expected according to sex, age, height and weight. In general analysis, we can consider that adolescents showed good quality of life; however, we considered the impairment of the *Pseudomonas aeruginosa* group as statistically significant in the evaluation of physical, respiratory, treatments, social/school, food, health and weight domains.

More studies evaluating adolescents with cystic fibrosis and associating the bacteriological profile of *Staphylococcus aureus* and *Pseudomonas aeruginosa* with respiratory muscle strength, quality of life and functional capacity are needed with greater sample size.

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